

JACC March 19, 2003

ABSTRACTS - Pediatric Cardiology 491A

1192-161

Comparison of Transthoracic Echocardiography and Cardiac Magnetic Resonance Imaging for Biventricular Function in Adult Patients After the Atrial Switch Procedure

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Background: Patients with transposition of the great arteries (TGA) and previous atrial switch procedure have a systemic right ventricle (RV). RV dysfunction frequently develops and requires serial life-long assessment. Cardiac magnetic resonance imaging (CMR) is emerging as the gold standard for RV imaging, but is not widely available. We examined whether echocardiographic variables of ventricular size and function were related to analogous measurements by CMR.

Methods: Eighteen stable adult patients in sinus rhythm (mean age 25.4 +/- 5.1 years) have been examined using both methods. Median age at operation was 1.8 years (range 2 months - 10 years). An HP 5500 Sonos ultrasound system was used for on line measurements in triplicate using 2D, M mode, and doppler images for RV and LV dimensions and function. CMR was performed using a 1.5 T Picker Edge or Siemens system. SPSS 10 for Windows was used for statistical analysis.

Results: Echocardiographic assessment of RV dimension (RV inlet relative to LV inlet size in the apical four chamber view) correlated strongly to CMR derived volumes (RV end diastolic volume (RVEDV) $r=0.92$; $p<0.001$, RV end systolic volume $r=0.66$; $p=0.039$) and mass (RV mass $r=0.81$, $p=0.008$). Echocardiographic measurements of right ventricular function (aortic ejection time and velocity time integral) correlated weakly with CMR measurement of RV ejection fraction (RVEF) ($r=0.5$; $p=0.042$; $r=0.46$; $p=0.053$, respectively). There was an inverse relationship between dP/dT and RVEDV ($r=-0.67$; $p=0.025$). Atrioventricular ring excursion at the septal wall by M-mode measurements correlated positively with CMR values of RVEF ($r=0.51$; $p=0.04$) and negatively with RV mass ($r=-0.614$, $p=0.044$). Echocardiographic left ventricular end diastolic dimension correlated well with CMR values of LV end diastolic volume ($r=0.64$; $p=0.026$).

Conclusion: Assessment of ventricular dimensions by echocardiography correlates well with CMR measurements in patients with previous atrial switch procedures for TGA and can be employed for serial assessment of the ventricles in this patient cohort.

1192-162

Right Ventricular Diastolic Function Before and After Pulmonary Valve Replacement Late After Repair of Tetralogy of Fallot Assessed by Cardiac Magnetic Resonance Imaging

Alexander van Straten, **Hubert W. Vliegen**, Mark G. Hazekamp, Hildo J. Lamb, Jaap Ottenkamp, Ernst E. Van der Wall, Albert De Roos, Leiden University Medical Center, Leiden, The Netherlands

Background: Pulmonary regurgitation (PR) in patients late after total correction for tetralogy of Fallot (TOF) may lead to progressive right ventricular (RV) systolic and diastolic dysfunction. Recently we assessed the beneficial effects of pulmonary valve replacement (PVR) on systolic RV function, however until now, the effects of PVR on diastolic function are still unclear. Purpose of this study was to assess diastolic RV function before and after PVR, using MRI.

Methods: Twentythree consecutive adult TOF patients who underwent PVR in our institution between 1998 and 2001 were studied. Median age at initial repair was 5.8 +/- 4.5 years (range 0.4 to 21.0) and median age at PVR was 31.2 +/- 9.4 years (range 17.0 to 45.6). Cardiac MRI was performed 6.1 +/- 3.4 months before and 20.3 +/- 3.3 months after PVR. Flow was measured through the pulmonary and tricuspid valve.

Results: Preoperative PR was 45% (range 25-64 %). At the second MRI, 6 patients (26%) had mild and one patient moderate PR (20-40%). Before PVR, 8 out of 23 patients showed end-diastolic forward flow in the main pulmonary artery (mean 10.3 +/- 3ml). After 20 months, only one patient had end-diastolic forward flow (EDFF) of 3ml ($P<0.01$). The E/A ratio as derived from the tricuspid flow curve, increased from 1.0 +/- 0.4 to 1.5 +/- 0.5 at 20 months post-PVR ($P<0.01$).

Conclusion: In adult patients late after total correction of TOF, PVR leads to a decrease of PR, a disappearance of EDFF and improvement of E/A ratio. These results indicate an overall improvement of RV diastolic function following PVR.

ORAL CONTRIBUTIONS

854 Clinical Pediatric Cardiology

Tuesday, April 01, 2003, 2:00 p.m.-3:30 p.m.

McCormick Place, Room S106

2:00 p.m.

854-1

Outcomes of Pediatric Acute Myocarditis

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Background: The optimum therapy for myocarditis in children is unknown. Design of trials of new therapies requires outcome data in the modern era in patients receiving steroids and/or intravenous immune globulin (IVIG). We present data on the outcomes of myocarditis in a large pediatric series using strict diagnostic criteria. **Methods:** Patients with myocarditis from 1985 to present were identified through a database search. Only those with biopsy-proven myocarditis or cardiac dysfunction and proof of concomitant

viral infection were included. Outcomes were defined as complete recovery (no residual dysfunction), incomplete recovery (continued dysfunction despite medical therapy) and death or transplantation. **Results:** There were 39 cases. 35 were biopsy-proven and 4 patients who were too unstable to undergo biopsy had proof of acute viral infection with pathogens known to cause myocarditis. 26 (67%) had complete recovery, 4 (10%) had incomplete recovery, and 9 (23%) either died (4) or underwent transplantation (5). Median time to death or transplant was 1.5 months (0.1 to 49 months). There were no late arrhythmias. 4 patients presented with complete heart block and normal function and had complete recovery without permanent pacemaker placement. 16 patients received steroids (10 complete recovery, 2 incomplete recovery, 2 deaths and 2 transplants), 1 received IVIG (complete recovery), 17 received steroids and IVIG (11 complete recovery, 2 incomplete recovery, 2 deaths and 2 transplants), and 5 received neither steroids nor IVIG (4 complete recovery, 1 transplant). Freedom from death or transplant was 81% at 1 year and 74 % at 5 years for all patients, with no difference between patients treated with steroids alone and those receiving additional IVIG. Median time to recovery of normal function in those treated with steroids alone was comparable to that of patients also treated with IVIG (1.8 and 2.0 months, respectively). **Conclusions:** We report contemporary outcomes of myocarditis in children. In this series, IVIG appeared to confer no advantage to steroid therapy alone. These data emphasize the need for randomized trials to assess the efficacy of current treatments as well as that of new therapies.

2:15 p.m.

854-2

Presenting Features and Outcomes for Children With Dilated Cardiomyopathy Who Develop Persisting Cardiac Dysfunction

Piers E. Daubenev, Alan W. Nugent, Patty Chondros, Michael Cheung, Andrew Davis, John B. Carlin, Steven D. Colan, Robert G. Weintraub, National Australian Childhood Cardiomyopathy Study, Melbourne, Australia

Background: A proportion of children with dilated cardiomyopathy (CM) develop persisting cardiac dysfunction. This study examines presenting features and outcomes for these children.

Methods: The National Australian Childhood Cardiomyopathy Study is a population-based study which includes all children within Australia with primary CM who presented at 0-10 years of age between 1987-1997. Diagnostic criteria for dilated CM comprised congestive heart failure with objective evidence of reduced cardiac function, a fractional shortening (FS) $\leq 20\%$ in those without symptoms, or pathological evidence of dilated CM. The prognostic factors sought included clinical features at presentation, results of all relevant investigations and serial echocardiographic parameters of LV function (expressed as Z scores). Late cardiac dysfunction was defined as persisting LV dilatation with a FS $\leq 20\%$ at two years after presentation. Study end-points were death or transplantation.

Results: See table. Of 24 children with late cardiac dysfunction, 11 (46%) died or underwent transplantation and another 11 (46%) have a dilated LV with continuing systolic dysfunction at latest follow-up.

Conclusion: Children with dilated CM who develop late cardiac dysfunction are characterized by LVH on their initial EKG, and greater LV dilatation with more depressed systolic function at 3 months after presentation. This group remains at risk of late death and the probability of ultimate recovery of ventricular function is low.

	Death/transplant within two years (n=52)	Late dysfunction (n=24)	Partial complete recovery (n=99)	P value
Median presenting age in months (range)	6.0 (0-120)	6.0 (.03- 111)	8.3 (0-112)	0.9 9
Positive family history	15 (29%)	4 (17%)	7 (7%)	0.0 02
Mean initial LVEDd Z (SD)	3.63 (2.81)	5.32 (2.96)	4.27 (2.45)	0.0 6
Mean initial LVFS Z (SD)	-11.58 (1.66)	-10.50 (2.56)	-9.80 (2.42)	0.0 005
Median(range) Z RV6 on initial EKG	0.84 (0-4.5)	1.9 (0.05- 6.6)	0.92 (0-4.6)	0.0 1
Mean LVEDdZat 3 months(SD)	5.83 (1.84)	5.26 (2.31)	2.85 (2.49)	<0. 000 1
Mean LVFS Z at 3 months (SD)	-10.58 (1.92)	-9.65 (2.98)	-5.31 (3.81)	<0. 000 1

2:30 p.m.

854-3

Tissue Doppler Predicts Mortality and Need for Transplantation in Children With Dilated Cardiomyopathy

Colin J. McMahon, Sherif Nagueh, Reanu Eapen, Irina Finkelstejn, Louis I. Bezold, Xiaoling Cao, Jack Price, William J. Dreyer, Jeffrey A. Towbin, Susan Denfield, Ricardo H. Pignatelli, Texas Children's Hospital, Houston, TX, Methodist Hospital, Houston, TX

Background: Characteristic tissue Doppler (TD) patterns and velocities have been determined in adults with various forms of cardiomyopathy. Limited data exists to identify children with dilated cardiomyopathy (DCM) who have a poor prognosis. This study sought to determine if TD predicts clinical outcomes in children with DCM.